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# PATHOLOGICAL CHANGES IN THE BRAIN IN CEREBRAL PALSY

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In spite of a number of investigations, cases of so-called cerebral palsy still present many problems in pathogenesis and etiology. However, according to neurological symptoms, cerebral palsy may be classified into several groups, and in the cases with similar symptoms the same parts of the brain are thought to be involved whatever the causal factors may be.

In our clinic, according to clinical symptoms, 331 cases of cerebral palsy were divided into four types; hypertonic, hypotonic, hyperkinetic, and mixed type. Out of 278 cases of the hypertonic type, 2 cases of low decerebration and 33 cases of high decerebration, in which the tonic neck reflexes of MAGNUS and de KLEYN and the righting reflexes were well developed, were found and the remaining cases were thought to be those of incomplete decerebration. Therefore, in these cases, pathological changes in the brain which cause the decerebrate state may be detected. This paper is a report on the pathological chansges in the brain in hydrocephalus and tuberculous meningitis, which were examined from the point of view mentioned above. These diseases form a part of causative factors of cerebral palsy as birth injuries do, and the study on the pathological changes caused by them may throw light on this field of neurology.

## MATERIALS AND STAINING METHODS

Pathological changes in the brain were examined in one case of hydrocephalus and seven cases of tuberculous meningitis.

The staining methods used were hematoxylin-cosin staining for the meninges and the neurons and WEIGERT technique for the nerve fibres.

## PATHOLOGICAL CHANGES IN HYDROCEPHALUS

The walls of both hemispheres are considerably thin, being one centimeter thick. However, the mesencephalon, pons, medulla oblongata and cerebellum appeared normal (Fig. 1). Microscopically, the cytoarchitecture of the both hemispheres, including the cerebral cortex, striate nuclei and thalamus, are completely

disappeared, the tissue is replaced by a large number of small glial elements (Fig. 2). The posterior part of the thalamus is saved from destruction, though a part of the neurons shows degenerative changes, such as shrinkage of the cell body, pyknosis or karyorhexis of the nucleus (Fig. 3). In the mesencephalon, pons, medulla oblongata and cerebellum, no pathological changes in the architecture are found (Fig. 4).

These lesions in the brain are equivalent to decerebration at the level of the thalamus.

## PATHOLOGICAL CHANGES IN TUBERCULOUS MENINGITIS

All seven cases show similiar pathological changes. Inflammatory processes of the meninges are most pronounced at the base of the brain about the optic chiasma and over the inferior surface of the pons, extending in a lesser measure upward over the cerebellum and the median and lateral surfaces of the cerebral hemispheres. The subarachnoid meshowrk over the base is infiltrated with mononuclear cells which are more plentiful immediately around the vessels. In addition, the basilar arteries, especially the striothalamic arteries, show characteristic changes, such as intimal proliferation and thrombus formation with resultant obliteration of most of these arteries (Fig. 5). These changes in the striothalamic arteries are thought to inevitably cause blockage of the blood supply to the striate nuclei and thalamus, which subsequently lead to necrosis. In reality, in these regions the neurons show an ischemic type of degeneration, manifested by shrinkage of the cell body, paling of the cytoplasm and pyknosis of the nucleus, and accompanied with reactive gliosis. In some cases, a complete dissolution of tissue about the vessels with scant glial reaction is found. In a striking contrast to these findings,

**Table**

| | | Hydrocephalus | | Tuberculous meningitis | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | 1 | | 2 | | 3 | | 4 | | 5 | | 6 | | 7 | | 8 | |
| | | l. | r. | l. | r. | l. | r. | l. | r. | l. | r. | l. | r. | l. | r. | l. | r. |
| Meninges | Base of brain | | | ++ | ++ | +++ | +++ | ++ | ++ | +++ | ++ | ++ | +++ | +++ | +++ | +++ | +++ |
| | Convex surface of hemisphere | | | + | + | + | + | + | + | + | + | + | + | ++ | ++ | + | + |
| | Surface of cerebellum | | | + | + | + | + | + | + | + | + | + | + | + | + | + | + |
| N. caudatus | | Completely | | + | +++ | +++ | +++ | + | ++ | ○ | +++ | +++ | +++ | +++ | ++ | +++ | +++ |
| Putamen | | Disappeared | | + | +++ | + | +++ | ++ | + | ○ | +++ | +++ | +++ | +++ | + | +++ | +++ |
| Globus pallidus | | | | + | + | +++ | +++ | ++ | + | ○ | +++ | +++ | ++ | + | + | +++ | +++ |
| Thalamus | | ++ | ++ | + | +++ | ++ | + | ○ | ○ | + | ○ | ± | ++ | + | ○ | +++ | +++ |
| N. ruber | | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ |
| Substantia nigra | | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ |
| Pons | | ○ | | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | ○ | + | ○ |
| Pyramidal tract | | | | +++ | + | ○ | + | ○ | ○ | + | + | ○ | + | ○ | ○ | + | + |

+++ marked lesion ++ moderate lesion + slight lesion ○ no lesion

the cerebral cortex shows only a slight necrotic change. A lesion of the pyramidal tract is absent or slight, except for one case. Moreover, the mesencephalon, pons, medulla oblongata and cerebellum are free of such lesion. These pathological changes in the brain are summarized in Table. Among these cases, Case 2 shows the following most typical pathological changes.

Fig. 6 shows the inflammatory changes in the meninges at the base of the brain. Perivascular infiltration with mononuclear cells is seen and the basilar arteries are boliterated by intimal proliferation or thrombus formation. However, these inflammatory processes are slight over the surfaces of the cerebral hemispheres and cerebellum (Figs. 7 and 8). In the striate nuclei and thalamus of the right hemisphere, blood vessels are scarcely found, and the architecture is evenly and diffusely obliterated, with only scattered shrunken neurons remaining, the tissue being replaced by glial elements (Figs. 9 and 10). In some parts the spongy structure due to loss of neurons and cyst formation about the shrunken vessel lacking blood cell are seen (Figs. 11 and 12).

The nerve fibres originating from these nuclei are considerably diminished, but the nerve fibres passing along the internal capsule remain uninvolved (Fig. 13).

In the striate nuclei and thalamus of the left hemisphere, a large number of capillaries filled with blood cells and neurons are found, though some neurons show a slight degenerative changes, such as paling and swelling of the cytoplasm. The nerve fibres orginating from these nuclei show hardly any appreciable pathological change. On the other hand, the majority of the nerve fibres passing along the internal capsule show an almost complete degeneration (Fig. 14). In the pons, though the transverse nerve fibres remain intact, the lesions of the pyramidal tracts, almost complete on the left and partial on the right side, are found (Fig. 15).

In short, the necrotic changes predominently involve the striate nuclei and thalamus in the territories of vascular supply by the striothalamic arteries which are obliterated by intimal proliferation and thrombus formation. On the contrary, the mesencephalon, pons, medulla oblongata andcerebellum are free of such necrotic change.

## COMMENT

According to Bieber and Fulton, bilateral decortication in a monkey (or mere removal of both frontal lobes) causes the upper extremities to assume a strongly semiflexed hemiplegic posture, and under this condition the reflexes of Magnus and de Kleyn are readily demonstrated. According to Sherrington, Wilson, Penfieldand Erickson, when a primate is decerebrated at the midcollicular level of Sherrington, thus excluding hypothalamus and rednucleus levels, a much more intense spastic rigidity develops and the upper extremities are thrust backwards in rigid extension with pronation, assuming the typical decerebrate attitude of Kinner Wilson. In this circumstance the rigidity is likely to be so intense that the Magnus and de Kleyn reflexes become difficult to bring out. The former procedure, decrotication

or removal of both frontal lobes, is designated "high decerebration" which is strictly differentiated from the latter, decerebration at the midcollicular level, designated as low decerebration.

In 2 cases out of our 331 cases, the upper extremities were thrust backwards in rigid extension with pronation, but the reflexes of MAGNUS and de KLEYN and righting reflxes did not develop. In 33 cases, the upper extremities assumed a semiflexed posture and the MAGNUS and de KLEYN reflexes and righting reflexes were readily demonstrated.

These cases of decerebration suggest that the cerebral hemisphere is the most favorite site of lesion, the parts of brain below the mesencephalon remaining uninvolved.

The pathological changes in hydrocephalus, such as the complete destructions of both cerebral hemisphers with intact mesencephalon, pons, medulla oblongata and cerebellum, are equivalent to high decerebration.

In cases of tuberculous meningitis, the lesions have a tendency to localize in the striate and thalamic region in the territories of vascular supply by the striothalamic arteries and less frequently to involve the pyramidal tract. These lesions in the brain also are equivalent to high decerebration, complete or incomplete.

MALAMUD (1950) reported on the pathological changes in 15 cases of cerebral palsy following either birth injury or inflammation of the central nervous system. In all cases characteristic marble state and precise bilateral localization in the striate area of the brain were found.

TOWBIN (1955) reported on the pathological changes in 23 cases of severe cerebral palsy. In his series, 6 cases with developmental disturbance of the cerebral cortex, 5 cases with cyst formation in the cerebral hemisphere, 1 case with lesion in the striate nuclei and internal capsule, 2 cases with lesion in the striate nuclei and thalamus, and 9 cases with atrophy of the cerebral hemisphere associated with marked enlargement of the lateral and third ventricles were found.

From these findings, it is probable that the lesions in the cerebral palsy have a tendency to localize predominently in the cerebral hemisphere whatever the causal factor may be. Moreover, these lesions in the brain must be the pathological changes causing the state of high decerebration, in which the MAGNUS and de KLEYN reflexes and righting reflexes develop.

## SUMMARY

1. In one case of hydrocephalus and seven cases of tuberculous meningitis, pathological changes in the brain were studied.

2. In hydrocephalus, a complete destruction of the both cerebral hemispheres except for the posterior part of the thalamus was demonstrated, and below the level of the corpora quadrigemina no lesion was found.

3. In tuberculous meningitis, the lesions were had a tendency to localize predominently in the striate nuclei and thalamus in the territories of vascular supply by the striothalamic arteries which were obliterated by inflammatory processes in

the basilar meninges. The lesion of the pyramidal tract was absent or slight except for one case. Below the level of the corpora quadrigemina, no appreciable change was detected.

4. These lesions in the brain are equivalent to high decerebration, in which the reflexes of MAGNUS and de KLEYN and righting reflexes develop.

Grateful acknowlegment I want to make to Prof. TETSUO ITO for his kind advice during this study.

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Explanations for the figures:

Plate I.

**Fig. 1.** Hydrocephalus. The walls of both hemispheres are considerably thin.

**Fig. 2.** Hydrocephalus. The tissue of the hemisphere are replacad by a large number of glial cells.

**Fig. 3.** Hydrocephalus. In the thalamus, many neurons are seen, though some of them show degenerative change.

**Fig. 4.** Hydrocephalus. In the corpora quadrigemina, most of the neurons show normal appearance.

**Fig. 5.** Tuberculous meningitis. The subarachnoid meshowork over the base is infiltrated with mononuclear cells which are more plentiful immediately around the vessels. The basilar artery shows intimal proliferation with resultant obliteration.

**Fig. 6.** Tuberculous meningitis (Case 2). Obliteration of the basilar artery due to intimal prliferation is seen.

Plate II.

**Fig. 7.** Tuberculous meningitis (Case 2). On the convex surface of the hemisphere, only a slight inflammatory process is found.

**Fig. 8.** Tuberculous meningitis (Case 2). On the convex surface of the cerebellum, a slight inflammatory process is seen.

**Fig. 9.** Tuberculous meningitis (Case 2). In the putamen, the neurons are almost completely disappeared, and replaced by glial cells.

**Fig. 10.** Tuberculous meningitis (Case 2). In the thalamus, the neurons are also considerably diminished, and replaced by glial cells.

**Fig. 11.** Tuberculous meningitis (Case 2). In some parts of the thalamus, spongy structure due to loss of neurons is seen.

**Fig. 12.** Tuberculous meningitis (Case 2). In other part of the thalamus, cyst formation about the shrunken vessel lacking blood cell is seen.

Plate III.

**Fig. 13.** Tuberculous menigitis (Case 2). In the right hemisphere, thalamus and striate nuclei

show marked lesion, but no appreciable degeneration of the nerve fibres passing along the internal capsule is found. Put., Putamen, Th., Thalamus.

**Fig. 14.** Tuberculous meningitis (Case 2). In the left hemisphere, thalamus and striate nuclei show no marked lesion, but the nerve fibres passing along the internal capsule are considerably diminished. Int. C., Internal Capsule.

**Fig. 15.** Tuberculous meningitis (Case 2). In the pons, though the trausverse nerve fibres remain intact, the lesions of the pyramidal tracts, complete on the left and partial on the right side, are found.

## 和文抄録

# 脳性小児麻痺の脳病変

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高　橋　哲　良

脳性麻痺の最大多数は痙直性麻痺であつて，その典型的な症例では高位除脳症状を呈し，Magnus and de Kleyn 緊張性頸反射や立直り反射が証明される．従つて脳性麻痺の脳病変として高位除脳に相当する病変があることが期待される．本報告に於ては内脳水腫1例，結核性脳膜炎7例，計8例に就いて脳病変を検査した．内脳水腫に於ては両側大脳半球が殆んど完全に破壊され，視床の後半部と，中脳，小脳，橋，延髄はよく保存されていた．この病変は典型的高位除脳の病変である．結核性脳膜炎7例に於ては，各症例共共通した病変が認められた．即ちすべて症例に於て炎症は脳底部脳膜に著しく高度であるが，大脳半球穹窿部や小脳の脳膜の炎症性変化は極めて軽微であつた．脳底部脳膜では激しい炎症の為に脳底動脈殊に A. striothalamicus も強く障害され，内膜増殖や血栓形式の為に閉鎖し，大脳核や視床えの血流障害を来し，その壊死を来していた．大脳皮質の障害は一般に軽微で，錐体路の破壊も比較的軽いものが多く，唯一例では殆んど完全に変性消失していた．これ等7例の症例の脳病変も高位除脳（完全又は不完全）に相当するものである．

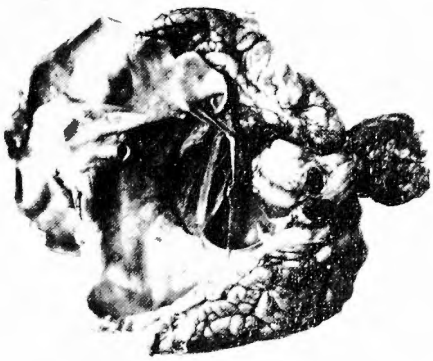


Fig. 1

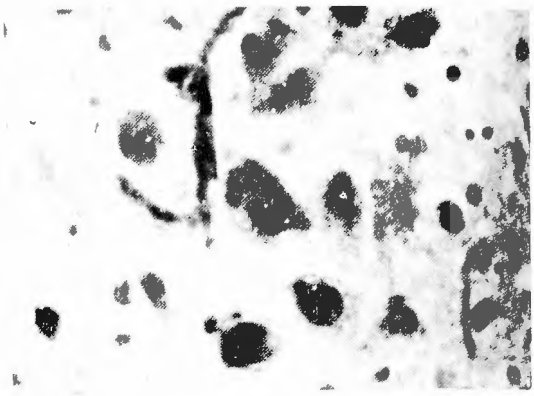


Fig. 4

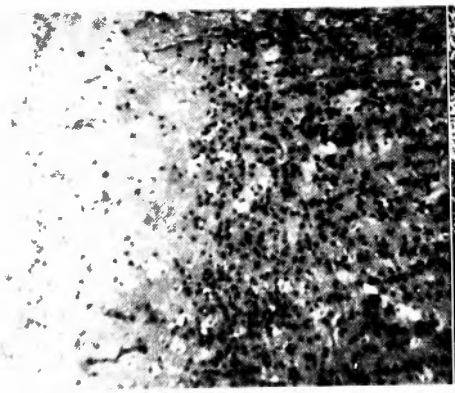


Fig. 2

Fig. 5

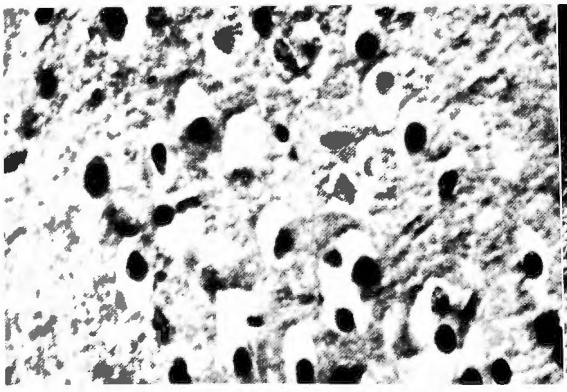


Fig. 3

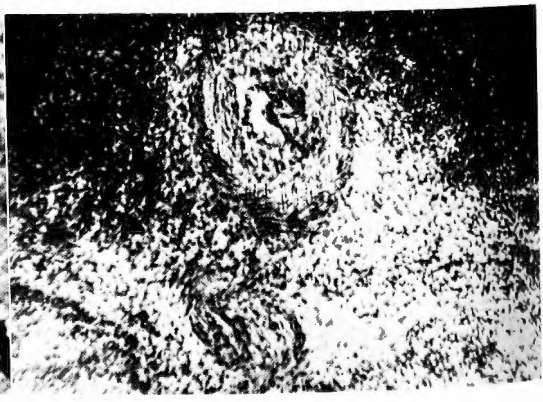
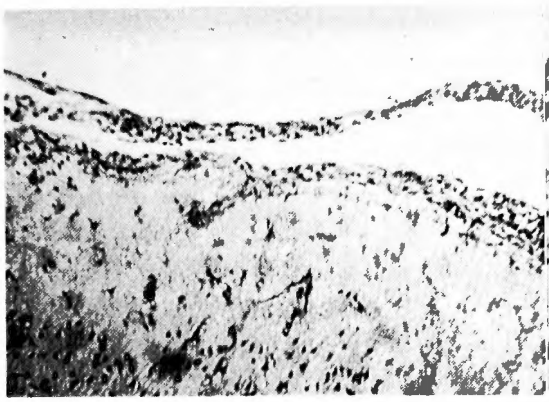


Fig. 6



Fgi. 7

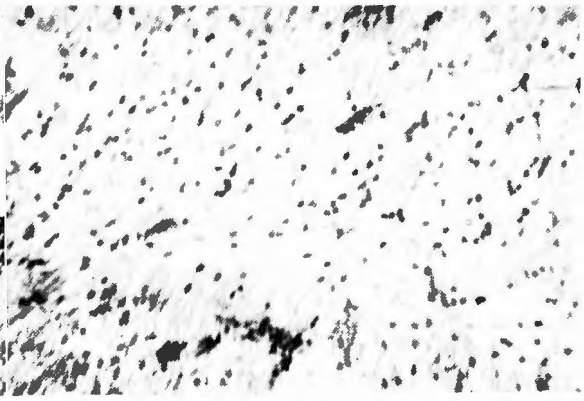


Fig. 10

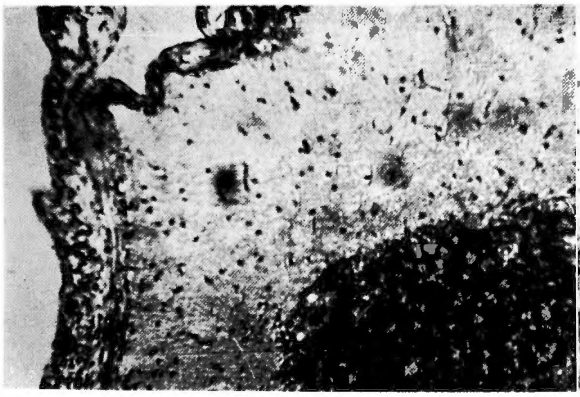


Fig. 8

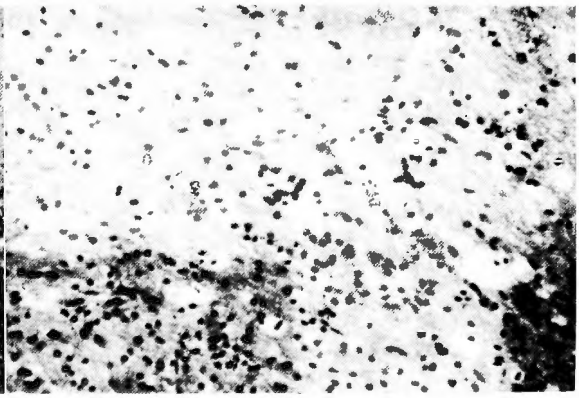


Fig. 11

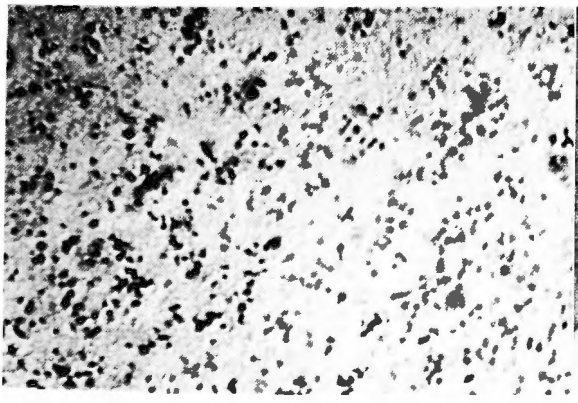


Fig. 9

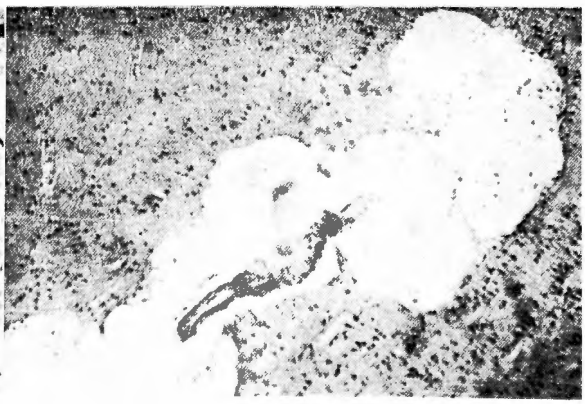


Fig. 12


Fig. 13


Fig. 14

Fig. 15